Can developmental disorders be used to bolster claims from evolutionary psychology? a neuroconstructivist approach

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Introduction

Data from adult neuropsychological patients and studies of individuals with genetic disorders are often used by evolutionary psychologists to motivate strong nativist claims about the organization of the neonate brain in terms of innately specified cognitive modules (Barkow, Cosmides & Tooby, 1992; Duchaine, Cosmides, & Tooby, 2001; Pinker, 1997). Such hypotheses are based, in our view, on static snapshots of phenotypic outcomes in middle childhood and adulthood and tend to ignore one vital causal factor affecting disorders, i.e., the actual process of ontogenetic development. In contrast to nativists, we take a truly developmental approach to both normal and atypical outcomes by focusing on the infant start state and the developmental trajectories that lead to such outcomes.

In this chapter, we discuss why it is essential to take a neuroconstructivist approach to interpreting the data from developmental disorders and why these latter cannot be used to bolster nativist claims. From our studies of older children and adults with the neurodevelopmental disorder, Williams syndrome, we show how processes that some claim to be "intact" actually display subtle impairments and cannot serve to divide the cognitive system into independent parts that develop normally from parts that develop atypically. Likewise, from our studies of infants and toddlers with developmental disorders, we identify low-level deficits in general capacities that have differential effects on the phenotypic outcome of different cognitive domains. Indeed, a tiny impairment very early in development can have a huge impact on some domains (the seemingly "selectively impaired cognitive modules") and a very subtle impact on other domains (the seemingly "intact cognitive modules"). It is thus crucial not only to focus on domains showing serious deficits in developmental disorders, but also to carry out in-depth studies of domains that seem at first blush to be unimpaired (Karmiloff-Smith, 1998). Because the brain develops as a whole system from embryogenesis onwards, we believe it to be highly unlikely that children with genetic disorders will end up with a patchwork of neatly segregated, preserved and impaired cognitive modules.

The above argumentation does not only hold for atypical development, of course. In keeping with some theorists of infant development, we also find it highly unlikely that the normal infant brain starts out with pre-specified modules solely dedicated to the independent processing of specific cognitive domains. Indeed, we challenge the

"Swiss Army Knife" metaphor adopted by some evolutionary psychologists for the neonate brain (Barkow et al., 1992; Duchaine et al., 2001). Rather, we argue that the infant brain is not like a Swiss Army knife simply handed down by evolution with pre-formed, specialised components that form, in the case of developmental disorders, a segregated pattern of individually impaired/preserved modules at birth. Rather, as Piaget did for the normal child (Piaget, 1953, 1971), we contend that ontogenetic development itself is the clue to understanding both normal and atypical development and its relation to the structure of the resulting adult cognitive system. In a similar vein to Piaget's constructivism (1953, 1971), we embrace the notion that the child constructs his own environment and sculpts the microcircuitry of his own brain through his physical and mental actions on the world.

How the infant brain sculpts itself through ontogenetic development

Undeniably, all constructs - including nativism - impute some role to the external stimuli. However, unlike staunch nativism that considers environmental stimuli as mere triggers to a genetic blueprint for development, and unlike staunch empiricism that sees the environment as the major contributor to cognitive outcomes, we contend that gene expression and environment constantly undergo complex and dynamic interactions that only an in-depth analysis of ontogenesis can reveal.

For example, the onset of complex functions in the cerebral cortex of the infant brain can be traced to a burst in synaptic activity– the formation of rich networks of

connections that allow knowledge to be encoded. This burst of synaptogenesis is under genetic control and appears to take place across the cortex relatively independently of input from the environment (Huttenlocher, 2002). However, synaptogenesis creates a surfeit of possible connections (many more than are retained in the eventual adult system), and it is the environment that selects which connections will be functionally useful. Unused connections are gradually eliminated. This pruning process continues over many years, i.e., well into adolescence for the frontal regions, for example, allowing the environment to shape the raw mechanisms that genetic processes have put in place. What is included in the notion of "environment"? First, for a given cognitive system within the organism, the "internal" environment potentially includes inputs from other cognitive systems as well as sensory inputs. "Environment" also includes the social and physical worlds external to the organism that provide a wide variety of inputs to the different sensory systems.

To reiterate, it is in our view highly improbable that the infant starts life with independently functioning cognitive modules, simply awaiting appropriate triggers from the environment. Rather, our argument is that infant brain development is an activity-dependent process in which the environment acts not merely as a trigger but actually plays an important role in sculpting the final outcome in terms of both structure and function. In our view, initial non-cognitive perceptual biases orient the infant towards certain aspects of the environment such as, for example, a sequential processor that pays particular attention to the flow of real-time speech output but less

attention to, say, static spatial inputs. With repeated exposure and repeated processing of certain types of inputs (such as speech in our above example), certain circuits of the brain become increasingly specialised (Elman, Bastes, Johnson, Karmiloff-Smith, Parisi & Plunkett, 1996; Johnson, 2001). Thus, a domain-relevant mechanism becomes a domain-specific mechanism as a function of development (Karmiloff-Smith, 1998). In other words, adult modules are, we contend, the result of a very progressive process of modularisation over developmental time (Karmiloff-Smith, 1992).

Neuroconstructivism

There are several competing theories about the structure of the infant brain at birth (see Johnson, 2001, for full discussion). Maturationists claim that different parts of the brain come on line sequentially during development as a result of genetic programming. They tend to explain the absence of a particular behaviour in infancy by the hitherto absence of functioning of a specific region of the brain. Interactionists, by contrast, claim that at birth most parts of the brain function to some degree, but that it is the network of interactions both within and across regions that changes as a function of development. We have termed this "neuroconstructivism" (Karmiloff-Smith, 1998) or more recently the "interactive specialisation approach" (Johnson, Halit, Grice & Karmiloff-Smith, 2002). Rather than waiting for a region to come on line maturationally, infant brain regions may initially be <u>more</u> active than the adult's until the processes of specialisation and localisation of function gradually stabilise. It has now been shown that seemingly identical overt behaviour in infants

and adults is supported by different brain regions or interactions between regions (e.g., Csibra, Davis, Spratling & Johnson, 2002; De Haan, Pascalis & Johnson, 2002; Neville, Mills & Lawson, 1992). By the time we reach adulthood, our brains are indeed highly structured and functionally specialised, but this in no way entails that we started out in infancy with anything like this structure in place.

A compelling example of very progressive specialisation and localisation comes from the development of infant face processing. What could be more evolutionarily important than species-specific recognition? If the nativist position held, then face processing would seem to be an ideal candidate for a built-in module, ready to function independently of other brain circuits as soon as appropriate triggering stimuli were presented. Yet, although a preference for face-like stimuli seems to be present from birth (Johnson & Morton, 1991), infant face processing is very different from adult face processing in terms of both behaviour and the brain circuits involved. Initially infants are just as likely to track pictures of real faces as those of very schematic faces with only three blobs in the appropriate eye and mouth regions. By 2 months of age, however, they only track real faces. But even the neonate preference is not constrained to face-like stimuli alone. Rather, the stimuli that are preferred are those that have more information in the upper region than the lower region, like a T-shape (Simion, Valenza, Macchi Cassia, Turati, & Umilta, 2002). While this happens to coincide with the overall visual stimulus of a face, it is clearly not an innately-given “face template”, the brain processing not being initially dedicated to face processing alone. So it seems that evolution does not need to provide more than a

domain-general kick-start to face processing, with the guarantee that the external environment will furnish massive face input early in life. After huge quantities of face inputs over the first months of life, even 6-month-olds do not display the brain activity typical of both 12-month-olds and adults in terms of binding the perceptual features of a facial stimulus (Csibra et al., 2002). It is also known that early on, both hemispheres of the infant brain actively process faces. However, by the end of the first year, processing of faces shifts predominantly to the right hemisphere, the one typically more active in older children and adults (de Haan et al., 2002).

These are but a few aspects of how face processing develops during infancy, highlighting the fact that it does not come ready to display adult-like functioning once face stimuli have triggered a so-called innately-specified module. On the contrary, infants seem to require hundreds of thousands of face stimuli to progressively develop their face processing expertise such that by the end of the first year of life they start to display adult-like processing in terms of both behaviour and underlying brain processes. We contend that any face processing module that ultimately exists in adults, and that could by then be selectively impaired (e.g., McNeil & Warrington, 1993), actually develops out of initial attention biases in interaction with the rich face processing experience available to the infant.

Further evidence for progressive neuroconstructivism comes from the study of infants with peri-natal unilateral brain lesions to the right hemisphere. A review of their subsequent face processing abilities between 5 and 14 years of age revealed two

things (de Haan, 2001). First, their impairments were mild compared to adults who had experienced similar damage – less than half the children exhibited impairments in face or object recognition compared to controls. Whatever the early damage, it had been attenuated by developmental plasticity. Second, face-processing deficits were no more common than problems identifying objects, and a face processing deficit never occurred in the absence of an object processing deficit. The specialization of face processing and its progressive separation from object processing appears to be purely a product of development, with the face recognition system emerging as a gradual specialization of an initially more general-purpose system. The dissociation of face and object recognition in the adult simply cannot be replicated by early damage to the normal system.

Now, nativists might claim that the progressive changes in infant face processing simply constitute the unfolding of a genetic timetable. However, other work on early processing of language, for example, challenges this. Neville and her colleagues examined the brain processes of toddlers when they listened to a series of words. They found that it was number of words that the infant could produce, and not maturational age, that predicted which brain circuits were used (Neville et al., 1992). In sum, the ball is in the court of the evolutionary psychologist to demonstrate that the infant brain is really anything like the metaphor suggested by the Swiss army knife with its highly specialized component parts in place from the outset.

A re-examination of data from developmental disorders

Adult neuropsychological patients may in some cases display highly specific impairments in their performance, suggesting independently functioning modules and impairment to a very specialised area of the brain. It must be recalled, however, that in the adult neuropsychological case, the adult has suffered a brain insult to a hitherto normally developed and highly structured brain. Such structure, as we have consistently argued, is the result of prior development and tells us nothing about the start state. Yet, at first blush, overt behavioural outcomes in older children and adults with genetic disorders seem also to present a neat case of preserved and impaired modules. So, why do we continue to question this? People with genetic disorders do not, in our view, have normal brains with parts preserved and parts impaired. Rather, they have developed an atypical brain throughout embryogenesis and subsequent postnatal growth, so we should expect fairly widespread impairments across the brain rather than a very localised one. How can we then reconcile our theoretical assumptions with the empirical data suggesting clear-cut selective impairments?

We argue that the empirical data themselves need to be re-examined, both from the viewpoint of the overt behaviour versus the underlying cognitive processes, and from the viewpoint of the control groups used to make theoretical claims about genetic disorders. To do this, we will take the example of one genetic disorder, Williams syndrome, and briefly examine three domains which some researchers have claimed to be "spared" in this clinical population: face processing, language and social cognition.

Williams syndrome

Williams syndrome is neurodevelopmental disorder caused by a submicroscopic deletion of some 24 genes on one copy of chromosome 7q.11.23 (Donnai & Karmiloff-Smith, 2000). It occurs in approximately 1 in 20,000 live births. Clinical features include several physical abnormalities that are accompanied by mild to moderate mental retardation and a specific personality profile. The interest of WS to neuroscience stems from its very uneven profile of cognitive abilities, with spatial and numerical cognition seriously impaired, while language, social interaction and face processing seem surprisingly proficient for a clinical population with IQs in the 50s to 60s range (Bellugi, Wang, & Jernigan, 1994; Udwin & Yule, 1991).

Work by Bellugi and her collaborators first drew attention to the potential theoretical interest of the seeming dissociations in the Williams syndrome cognitive phenotype (Bellugi, Marks, Bihrle & Sabo, 1988). Surprising proficiency with language was shown to co-exist with serious problems with non-verbal tasks, in particular those calling on spatial processing. People with WS scored at floor, for example, on the Benton Line Orientation Task, but were within the normal range on the Benton Face Processing Task (Bellugi et al., 1988). This striking contrast between facial and spatial processing led some researchers (e.g., Bellugi et al., 1988) to maintain that face processing in WS is “intact” demonstrating, together with prosopagnosia (the inability to identify previous known faces) in the adult neuropsychological patients, that face processing is an independently functioning module.

Face processing in Williams syndrome

The early claims about an intact face processing module in WS have since been challenged, not with respect to the behavioural data themselves, but targeting the underlying cognitive and brain processes involved. Several studies have now replicated Bellugi's findings showing indeed that older children and adults with WS achieve behavioural scores in the normal range on some face processing tasks (Grice, Spratling, Karmiloff-Smith, Halit, Csibra, de Haan, & Johnson et al., 2001; Karmiloff-Smith, 1997; Udwin & Yule, 1991). However, this behavioural success is only superficially the same as that of normal controls. Usually we process faces configurally; our brains rapidly analyse the spatial relations between facial elements. By contrast, people with WS tend to predominantly analyse faces featurally: they focus more on the separate elements of a face, and less on the relations between elements (Deruelle, Mancini, Livet, Casse-Perrot, & de Schonen, 1999; Karmiloff-Smith, 1997; Rossen, Bihrle, Klima, Bellugi & Jones, 1996). So the cognitive processes underpinning the superficially successful face processing of people with WS seem to be different from the normal case.

A similar situation holds for the electrophysiology of the brain (Mills, Alvarez, St. George, Appelbaum, Bellugi, & Neville, 2000; Grice et al., 2001, in press). People with WS are more likely to show a predominance of the left hemisphere when processing faces in contrast to the typical right hemisphere dominance for face processing. Furthermore, people with WS do not display the normal inversion effect,

whereby upside down faces are processed differently from upright faces. In WS, both types of display are processed in the same way, again suggesting that this clinical group processes predominantly all face stimuli on a feature-by-feature basis. This cognitive difference does not hold only for facial stimuli. Work by Deruelle and her collaborators revealed that people with WS are more inclined to use featural than configural processing also of non-face displays (Deruelle et al., 1999). In sum, people with WS do not present with a normally developed "intact" face processing module and an impaired space-processing module, as nativists would claim. Rather, from the outset they have followed an atypical developmental trajectory such that both facial and spatial processing reveal a similar underlying impairment in configural processing. It is simply because the problem space of face processing lends itself more readily to featural analysis than spatial analysis does, so that it merely seems normal in the older child and adult. In other words, a fairly low-level impairment in configural processing early on impacts differentially on face processing and space processing during development, such that one domain can call on certain compensatory processes whereas the other cannot.

Language in Williams syndrome

Perhaps face processing just happens not to be the right domain for the evolutionary psychologist to establish a dissociation between innate components of the cognitive system. So, let's briefly examine another domain. Early claims were made for another dissociation in WS, this time between language and cognition. Language has been argued to be an innate mental organ specific to humans, and not reliant on general cognition (Pinker, 1994). So, on this account, we might expect certain genetic

disorders to allow normal language to develop even in the presence of an impairment to general cognition. Such a dissociation was initially claimed for WS. But as we have seen, such dissociations are actually highly unlikely given what we know about the processes of language development. And, in exactly the same way as our example from face processing, subsequent careful analysis of the ostensibly "intact" language capacity in WS revealed many, sometimes quite subtle, atypicalities which suggested that WS language was learnt via an atypical developmental trajectory (Karmiloff-Smith, et al., 1997; Laing, Butterworth, Ansari, Gsodl, Longhi, Panagiotaki, Paterson, & Karmiloff-Smith, 2002; Nazzi & Karmiloff-Smith, 2002; Nazzi, Paterson & Karmiloff-Smith, 2002; Singer-Harris, Bellugi, Bates, Jones & Rossen, 1997; Vicari, Brizzolara, Carlesimo, Pezzini, & Volterra, 1996; Volterra, Capirci, Pezzini, Sabbadini, & Vicari, 1996).

Initial comparisons were made between the abilities of individuals with WS and those from other syndromes who present with equivalent general cognitive abilities. Certainly compared to a disorder such as Down syndrome (DS), language in WS appears strikingly more advanced. For example, while the language of individuals with DS often shows appropriate word ordering, their speech remains telegraphic, with a reduced use of function words, poorly inflected verbs, predominant use of the present tense and a lack of appropriate feature marking on pronouns and anaphors, a state that largely persists into the adult years (Fowler, Gelman & Gleitman, 1994). On the other hand, the language of individuals with WS often reveals sophisticated

linguistic knowledge. For instance, in an analysis of the expressive language of four children with WS, Clahsen and Almazan (1998) reported the presence of complex syntactic structures and grammatical morphemes that were almost always used correctly.

A number of studies have pursued comparisons between language in WS and DS, presumably under the view that DS can serve as a baseline of what one might expect of language development in the presence of mental retardation, against which the achievements of WS may be measured (see discussion in Karmiloff-Smith, Ansari, Campbell, Scerif, & Thomas, in press). Thereafter, however, detailed investigations began to demonstrate that language performance is not at normal levels in WS, and at the very least shows a developmental delay of at least two years (Singer-Harris, et al., 1997). Most recent studies that compare the performance of individuals with WS to typically developing children now use a control group matched for mental age, to which their performance levels are more closely tied. Paradoxically, this matching procedure implicitly concedes that language development in WS is not independent of general cognitive ability!

While the language performance of individuals with WS is relatively impressive (compared to other syndromes with low IQs), evidence of atypicalities has accumulated in all areas of language, and at all stages of language development,

including vocabulary, syntax, morphology, and pragmatics, as well as the precursors to language development in infants (see Thomas & Karmiloff-Smith, 2003, for a review). Moreover, comparisons with Down syndrome actually exaggerate the apparent language ability in WS, given that individuals with DS demonstrate a particular developmental deficit in phonological processing which is not found in WS. And, most crucially, when this pattern of deficits in the endstate of each disorder – better language in WS than DS – was traced back to the respective abilities in early language comprehension in infancy, the pattern had disappeared. Infants with WS and DS showed equal (and very delayed) early language comprehension, implying that adult phenotypes were the product of differential atypical trajectories of development (Paterson. Brown, Gsodl, Johnson, & Karmiloff-Smith, 1999).

Social cognition in Williams syndrome

The story we have seen for face recognition and for language development in WS is now being repeated in the study of social cognition in this disorder. Here again, an initial claim was made that in WS, social cognition developed normally against a background of other impaired functions. Yet, here again, subsequent detailed research has suggested that social cognition and pragmatics are atypical in WS, sometimes subtly but sometimes quite markedly (Tager-Flusberg & Sullivan, 2000; Jones, Bellugi, Lai, Chiles, Reilly, Lincoln & Ralphs, 2000). The study of WS illustrates that in every case a ‘preserved function’ has been heralded in this genetic developmental disorder, it did not stand up to subsequent detailed investigation. Indeed, whenever a claim has been put forward that is inconsistent with what we

know about development in general, this claim has turned out to be false. And similar results have also start to emerge from other developmental disorders with a genetic basis, such as in the study of Specific Language Impairment, developmental dyslexia, Fragile X syndrome and Velo-cardiof3acial syndrome (see discussions in Chiat, 2001; Karmiloff-Smith, 1998; Karmiloff-Smith, et al., in press; Thomas, 2003; Thomas & Karmiloff-Smith, 2002).

The importance of neuroconstructivism

What becomes clear from the above examples is that genetic disorders do not provide data pointing to neatly impaired and spared cognitive domains that lend themselves to the evolutionary psychology claims. Rather, studies of developmental disorders demonstrate just how very complex and dynamic are the processes of gradual ontogenetic development and how important it is to recall that for humans, selection has favoured a very lengthy period of postnatal brain development. It is one thing to spot consistency in the pattern of adult cognitive structures following development in the environments to which human adults are typically exposed. It is quite another, however, to assume - against accumulating counter-evidence - that these structures are innately present in the infant brain. And it is yet a further act of faith to then argue that selection somehow favoured them!

So, what's wrong with selection, one might ask! Have its mechanisms gone awry? Why is a process as crucial to recognising conspecifics as, say, face processing not innately specified and cordoned off to function independently from all other

processes? The reason may well lie in two different types of control, and the fact that some higher-level cognitive outcomes may not even be possible at all without the gradual ontogenetic process of learning (Elman et al., 1996; Piaget, 1971).

It is generally accepted that there are two forms of biological control: mosaic control and regulatory control (Elman et al., 1996). Mosaic control involves deterministic epigenesis: genes tightly control timing and outcome, the process is fast and operates independently of other processes. This form of control is fine under optimal conditions. However, it places serious limits on complexity and flexibility of the developmental process. Some parts of human development are likely to involve mosaic control, such as the very basic macrostructures of the brain and of the body. However, the other type of control, regulatory control, is much more common and involves probabilistic epigenesis. It is especially prominent in the developing microstructure of the brain. It is under broad rather than tight genetic control, is slow and progressive, with limited pre-specification. In this type of control, different parts of a system develop interdependently. And, unlike in mosaic control, there are fewer constraints on complexity and plasticity. This does not mean, of course, that there are no biological constraints, as the empiricist position might claim, but it is far less constrained than mosaic control. Genes and their products are most unlikely to code for the cognitive level, but rather for differences in developmental timing, neuronal density, neuronal migration, neuronal type, firing thresholds, neurotransmitter differences and the like.

The notion of neuroconstructivism embodies regulatory control, with ontogeny seen as the prime force for turning a number of domain-relevant learning mechanisms progressively into domain-specific outcomes in the adult. This does not imply that the infant brain is a single, homogeneous learning device – there is, no doubt, much heterogeneity in the initial gross wiring of the brain. But this heterogeneity bears little resemblance to the ultimate functional structures that can only emerge through the process of ontogeny. In other words, rather than the mosaic form of tight genetic control which some evolutionary psychology models invoke, the human brain may well have evolved to favour very progressive development and neuroconstructivist plasticity rather than prespecification. If we are to understand what it is to be human, our continuing emphasis must be on the process of development itself.

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